Case Report

Wegener's Granulomatosis Presenting as Wallenberg Syndrome: A Case Report

Maziar Emamikhah, MD,* Farzad Sina, MD,* Mahisa Mokhtari, MD,* Fatemeh Shirani, MD,† and Mohammadreza Asadipanah, MD‡

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, a vasculitis affecting small and medium sized vessels usually affects the upper and lower respiratory tract, the kidneys, and the eyes. Neurologic manifestation in central nervous system (CNS) is less frequent than the peripheral and usually is in form of stroke. Few cases of lateral medullary ischemic stroke (Wallenberg syndrome) due to GPA have been reported. A 41 year-old female, presented with acute vertigo, nausea/vomiting, hiccups, dysphagia. In physical examination she had a saddled nose, horner syndrome, soft palate paralysis, crossed hypoesthesia of face, and limbs and hemi-ataxia. Brain magnetic imaging revealed a left lateral medullary infarction and sinusitis confirmed by paranasal Sinus CT scans. Chest CT showed a cavitary mass. Laboratory findings were remarkable for anemia, elevated erythrocyte sedimentation rate, and C-reactive protein. Cytoplasmic antineutrophil cytoplasmic antibody (ANCA)/anti-proteinase 3 was positive. Diagnosis of GPA was established and treatment was started. During 6-month follow-up improvement was satisfying and no relapses occurred. Medullary infarct is reported in few GPA patients, especially at presentation. Definite diagnosis is based on tissue biopsy. Although in context of extra CNS involvement and positive ANCA diagnosis can be made confidently. Treatment of choice in CNS involvement is not clear, corticosteroids and immunosuppressives seem effective. CNS involvement, especially stroke may present GPA or accompany extra CNS symptoms. Prompt diagnosis and treatment is essential.

Key Words: Granulomatosis with polyangiitis—granulomatosis—Wegener—lateral medullary syndrome—Wallenberg syndrome—stroke

Introduction

Granulomatosis with polyangiitis (GPA), formerly Wegener's granulomatosis, is a necrotizing granulomatous vasculitis of small-medium sized vessels.[1-4] It is an antineutrophil cytoplasmic antibody (ANCA) – associated vasculitides.[4] GPA affects upper (sinusitis) and lower respiratory tracts (nodules, etc), the kidneys ,and the eyes.[1] Half of patients manifest neurologic involvement, mainly as peripheral nerve disorders. Less common central nervous system (CNS) involvement is in 2 forms, granulomatous (G-CNS) and vasculitic (V-CNS).[1-3] Latter often, is an ischemic or hemorrhagic (rarer) stroke.[1-3,5] Cerebritis, encephalopathy,[2,6] subarachnoid hemorrhages, ischemic myelopathy, and arterial/venous thrombosis[2,7] are rarely reported forms. Among

From the *Department of neurology, Rasool-e Akram hospital, Iran University of Medical Sciences, Tehran, Iran; †Department of rheumatology, Rasool-e Akram hospital, Iran University of Medical Sciences, Tehran, Iran; and ‡Department of internal madicine, Rasool-e Akram hospital, Iran University of Medical Sciences, Tehran, Iran.

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Address correspondence to Maziar Emamikhah, MD, Neurologist, Department of neurology, Rasool-e Akram Hospital, Iran university of Medical Sciences, Niyayesh street, Sattarkhan, Tehran, Iran. E-mail address: maziar.emamikhah@yahoo.com.

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strokes, Wallenberg syndrome (WS) has been observed in few.[1-3,8-10] Much scarcer is WS as presenting symptom.[1-3] Here we report such a patient.

Case Report

A 41-year-old female, without any prior known medical conditions, presented with acute vertigo, nausea, vomiting, hiccups, and dysphagia. Examination revealed a young woman with a mildly saddled nose, left horner syndrome (partial ptosis, miosis, and anhidrosis), soft palate paralysis, hemi-ataxia, and crossed hypoesthesia (Left face, right extremities). A stroke, WS, was presumed clinically. Brain magnetic imaging revealed left lateral medullary infarction and marked sinusitis, confirmed by paranasal-SinusCT-scan (Fig 1).

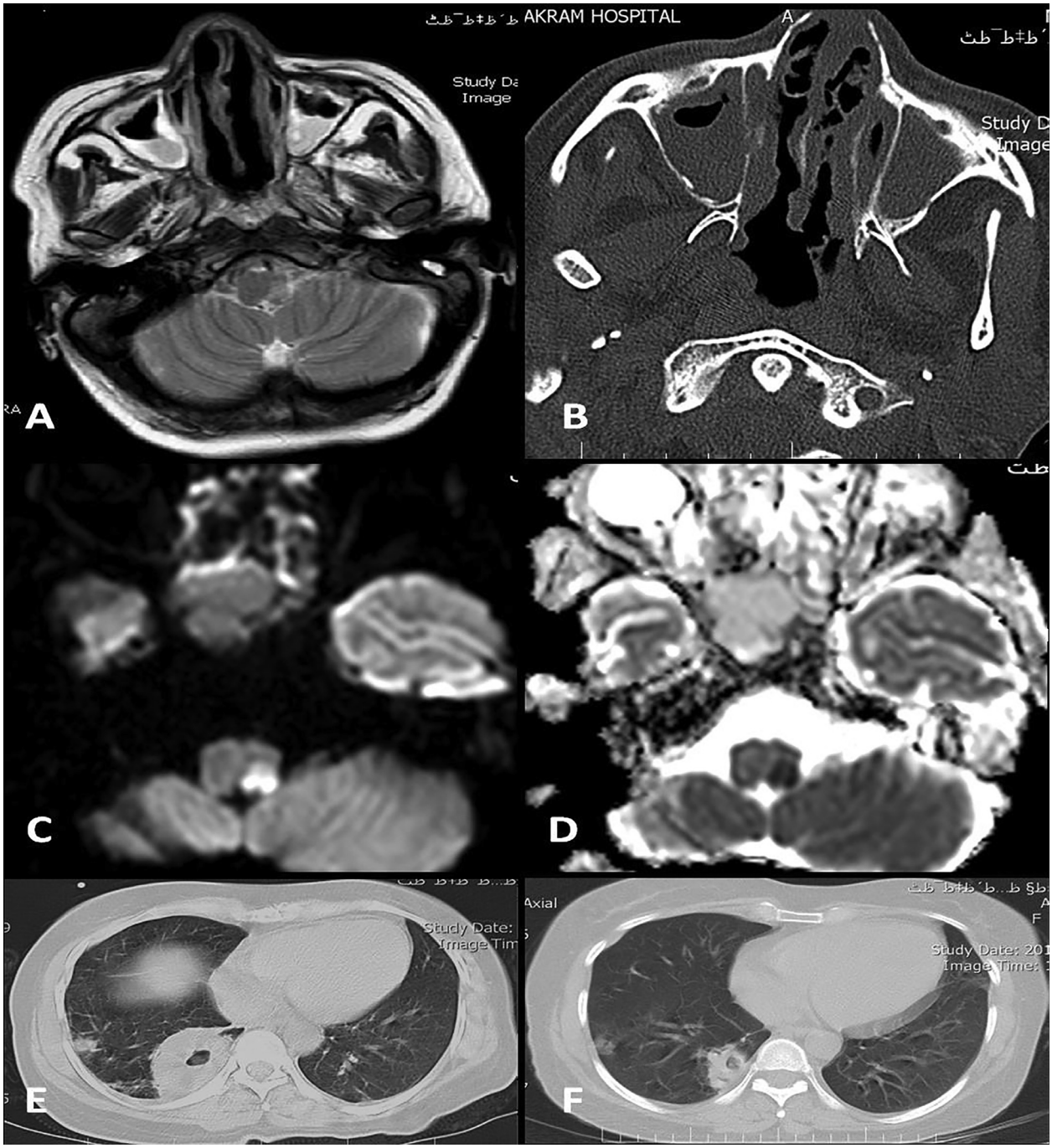

Figure 1. *(A) Brain MRI, T2-hypersignal area in left lateral medulla and mucosal thickening and fullness of maxillary sinuses. (B) PNS-CT, severe sinusitis in paranasal sinuses. (C and D) Diffusion imaging restriction in favor of acute stroke. (E and F) Chest CT shows bilateral infiltrations and a mass-like lesion with cavitation in right lung before (left plane) and 6 month after (right plane) treatment. Abbreviation: MRI, magnetic resonance imaging; PNS-CT, paranasal sinuses computed tomography.*

Echocardiography was normal. Comprehensive brain and cervical Doppler ultrasound and brain MR-Angiography results were negligible for any abnormalities in cervical and brain arteries including vertebral arteries. During admission coughs appeared and chest CT revealed a cavitary mass (Fig 1). Laboratory findings were remarkable for anemia (Hb = 8), elevated erythrocyte sedimentation rate (ESR = 90) and C-reactive protein (CRP = 96), transient elevated aminotransferases and creatinin.

Considering above findings a vasculitis (most probably Wegener's granulomatosis) was assumed. High titers of cytoplasmic ANCA/anti-proteinase3 (C-ANCA/PR3 = 100, negative <12 U/mL) established the diagnosis. High-dose methylprednisolone, cyclophosphamide, and aspirin started. After improvement patient was discharged receiving high-dose prednisolone and monthly cyclophosphamide. During 6-month follow-up, symptoms, erythrocyte sedimentation rate, CRP, and chest-CT (Fig 1) improved and no relapses occurred.

Discussion

Our patient is unique, because the presentation leading to GPA diagnosis was a rare form of V-CNS and extra-CNS involvements remained quite asymptomatic. Definite diagnosis of V-CNS is based on tissue biopsy. Although in context of extra-CNS involvement and positive ANCA (≈90% of cases) diagnosis can be made confidently.[1-3] Treatment of choice in V-CNS is not clear, but based on few reports induction with high-dose corticosteroids combining with cyclophosphamide or Rituximab seems practical and effective, as was in our patient. Maintenance therapy with prednisone, plus Azathioprin, Methotrexate, Mycophenolate, Rituximab, or infliximab has been successful.

Conclusion

CNS involvement (e.g. stroke) may present GPA before extra-CNS symptoms. Prompt diagnosis and treatment, is essential to prevent further sequels and warranting favorable outcome.

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